

Association Between Decline in Slow Vital Capacity and Respiratory Insufficiency, Use of Assisted Ventilation, Tracheostomy, or Death in Patients With Amyotrophic Lateral Sclerosis.

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ALS患者における、Slow Vital Capacityと呼吸不全、人工呼吸器の使用、気管切開、死亡リスクの関連。

Table 1. Baseline Demographics and Disease Characteristics¹⁸⁻²⁰

Variable	EMPOWER Placebo Group (n = 456)	BENEFIT-ALS Placebo Group (n = 210)	PRO-ACT Database Placebo Patients (n = 227)	Total (N = 893)
Age, mean (SD), y	57.3 (11.3)	56.8 (10.6)	55.4 (11.6)	56.7 (11.2)
Sex, No. (%)				
Male	289 (63.4)	148 (70.5)	148 (65.2)	585 (65.5)
Female	167 (36.6)	62 (29.5)	79 (34.8)	308 (34.5)
BMI, mean (SD)	26.1 (4.3)	26.8 (4.4)	25.3 (4.0)	26.1 (4.3)
White, No. (%)	428 (93.9)	175 (83.3)	218 (96.0)	821 (91.9)
Duration of symptoms, mean (SD), mo	15.6 (5.4)	26.7 (23.7)	18.6 (8.3)	19.0 (13.6)
Time from diagnosis to baseline, mean (SD), mo	7.7 (5.0)	12.2 (17.1)	8.3 (6.9)	8.9 (9.8)
Family history of ALS, No. (%)	24 (5.3)	12 (5.7)	ND	ND
Riluzole use, No. (%)				
No	115 (25.2)	73 (34.8)	ND	ND
Yes	341 (74.8)	137 (65.2)	ND	ND
Baseline SVC, mean (SD)	89.3 (17.7)	89.7 (17.2)	93.5 (15.6)	90.5 (17.1)
Baseline SNIP, mean (SD)	55.0 (26.3)	61.4 (25.7)	ND	ND
Site of ALS symptom onset, No. (%) ^a				
Bulbar	111 (24.3)	30 (14.3)	42 (18.5)	183 (20.5)
Other	345 (75.7)	180 (85.7)	187 (82.4)	712 (79.7)
Baseline ALSFRS-R total score, mean (SD)	38.0 (5.6)	37.3 (4.2)	ND	ND

Abbreviations: ALS, amyotrophic lateral sclerosis; ALSFRS-R, Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised; BMI, body mass index (calculated as weight in kilograms divided by height in meters squared); ND, no data; SNIP, sniff nasal inspiratory pressure; SVC, slow vital capacity.

^a Two patients in the PRO-ACT database reported both bulbar and other for site of ALS symptom onset.

Table 3. Reduction in Risk of Amyotrophic Lateral Sclerosis Milestones With Decrease in the Slope of Decline in Percentage Predicted Slow Vital Capacity by 1.5 Percentage Points per Month^a

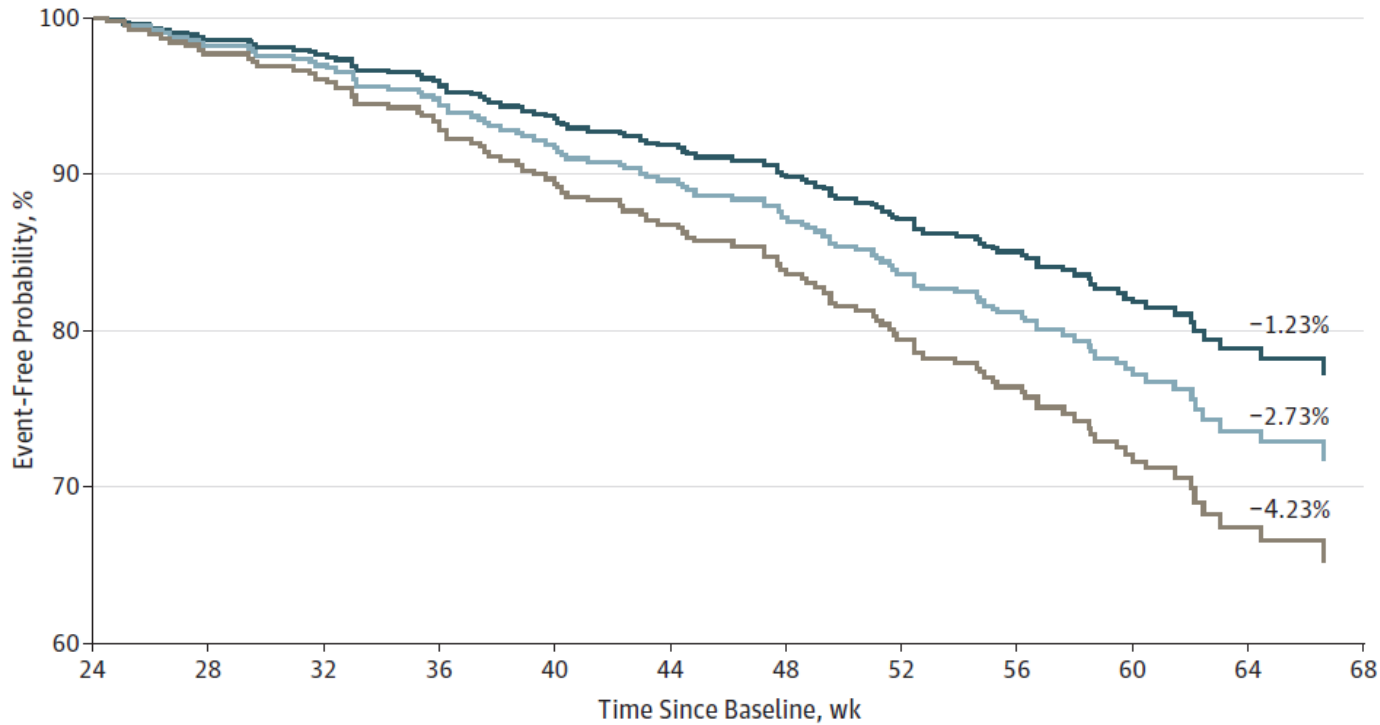
Variable	Risk Reduction, % (95% CI)
Decline in the ALSFRS-R subdomain or death after 6 mo	19 (12-25)
First onset of respiratory insufficiency or death after 6 mo	22 (18-27)
First occurrence of tracheostomy or death after 6 mo	23 (19-27)
Death at any time after 6 mo	23 (18-27)

Abbreviation: ALSFRS-R, Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised.

^a Data are based on a Cox proportional hazards regression model of time to clinical events from the month 6 visit to end of the follow-up period ($P < .001$ for all comparisons).

1.5%/月のSVCの低下率を回避すると、呼吸に関連するイベントや死亡リスクを19-23%回避できる。

Figure 2. Probability of Respiratory Failure-Free Survival Related to the Rate of Decline of Slow Vital Capacity



No. at risk	24	28	32	36	40	44	48	52	56	60	64	68
-1.23%	441	433	427	419	406	397	388	374	354	238	134	78
-2.73%	441	432	425	414	399	388	375	359	337	220	115	59
-4.23%	441	431	421	409	390	377	361	342	318	198	92	35

Data are based on a Cox proportional hazards regression model of time to clinical events. The 3 percentages are slope of change in percent predicted slow vital capacity per month.

SVCの低下率が大きいと、呼吸に関連するイベントや死亡率が高い。